General

Guideline Title

Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition).

Bibliographic Source(s)

RHDAustralia (ARF/RHD writing group), National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand. Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). Casuarina (Australia): RHDAustralia; 2012. 134 p. [459 references]

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: ARF/RHD Guideline Development Working Group of the National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand. Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia: an evidence-based review. Sydney (Australia): National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand; 2006 Jun. 84 p.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- March 22, 2016 – Opioid pain medicines : The U.S. Food and Drug Administration (FDA) is warning about several safety issues with the entire class of opioid pain medicines. These safety risks are potentially harmful interactions with numerous other medications, problems with the adrenal glands, and decreased sex hormone levels. They are requiring changes to the labels of all opioid drugs to warn about these risks.

Recommendations

Major Recommendations

The levels of evidence (I-IV) and grades of recommendations (A-D) are defined at the end of the "Major Recommendations" field.

Primordial and Primary Prevention of Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease (RHD)

Recommendations Regarding the Primordial and Primary Prevention of ARF and RHD

Primordial Prevention

While there is only limited evidence to support the effectiveness of specific initiatives in the primordial prevention of ARF/RHD, ecological data would suggest that the risk of ARF/RHD is linked to poverty and disadvantage. Housing and overcrowding would appear to be one important factor. However, given the uncertainties regarding specific causes, advocating for the primordial prevention of ARF/RHD, based on one or another specific environmental or social strategy, cannot be supported. The broader context of equity, poverty alleviation and justice, in association with the empirical link observed between improved socioeconomic and environmental factors and reduced ARF incidence, as well as many other health conditions, should be sufficient to drive advocacy and change.

Primary Prevention

Primary prevention measures aimed at preventing ARF/RHD through the prevention or eradication of pharyngeal group A streptococcus (GAS) colonisation, or the early identification and treatment of GAS pharyngitis, are of uncertain effectiveness. While programs aimed at preventing GAS colonisation through antibiotic use may be effective in the short term, any long-term implementation is likely to be unsustainable, due to prohibitive costs, client inconvenience and the risk of antibiotic resistance. A GAS vaccine offers the possibility of a longer-term solution. While significant hurdles remain in the development of a safe, effective and affordable vaccine that can be provided to populations at highest risk of ARF/RHD, this should remain a priority.

Although some programs aimed at the identification and treatment of GAS colonisation have shown promise, the evidence supporting such an approach remains poor. In line with preventing GAS colonisation, such initiatives are also likely to be unsustainable, due to cost, client inconvenience and the risk of antibiotic resistance. Although the cost of managing established RHD is high, the number needed to treat to prevent RHD through such primary prevention programs would be high.

While the early treatment of GAS pharyngitis in highly-controlled environments (e.g., military camps) can prevent the subsequent development of ARF, there is no evidence that community-based programs that focus on the early treatment of GAS pharyngitis are effective in reducing the risk of ARF. The treatment of pharyngitis, as part of comprehensive and accessible primary healthcare, remains important. In this context, the education of patients, carers, schools and communities is crucial to ensure that the detection of symptomatic pharyngitis prompts primary healthcare attendance.

The utility of clinical scoring systems or rapid antigen detection tests (RADT) is variable in differentiating GAS and non-GAS pharyngitis. The development and validation of these and newer rapid diagnostic tests in Aboriginal and Torres Strait Islander populations at risk of ARF/RHD should be a priority. Empirical treatment of all cases of pharyngitis or throat swab-directed treatment should remain the priority in populations at high risk of ARF. The lack of a clear episode of symptomatic pharyngitis in all people presenting with ARF will mean there is an inherent failure rate in even the most comprehensive GAS pharyngitis treatment programs.

The link between skin-related GAS infection and the pathogenesis of ARF/RHD remains contentious. The role of GAS skin infection treatment in the primary prevention of ARF/RHD remains unproven, and is likely to be unsustainable without addressing the underlying causes of skin disease (see 'Primordial Prevention', above). Nonetheless, as with pharyngitis, the management of skin disease should remain a component of high-quality, comprehensive and accessible primary healthcare for all populations, irrespective of ARF/RHD risk.

Diagnosis and Management of Acute Rheumatic Fever

See Table 3.2 in the original guideline document for updated Australian guidelines for the diagnosis of acute rheumatic fever.

Clinical Features of Acute Rheumatic Fever: Major Manifestations

Arthritis

Because of the migratory and evanescent nature of the arthritis, a definite history of arthritis, rather than documentation by the clinician, is sufficient to satisfy this criterion (Grade D).

In high-risk populations in Australia, mono-arthritis or polyarthralgia are a common manifestation of ARF, and are often associated with overt or subclinical carditis. In these populations, aseptic mono-arthritis or polyarthralgia may be considered as a major manifestation, in place of polyarthritis (level IV, Grade C).

Sydenham's Chorea

Echocardiography is essential for the assessment of all patients with chorea, regardless of the presence of cardiac murmurs (level IV, Grade C). A finding of subclinical carditis is sufficient to confirm the diagnosis of ARF in high-risk populations (Grade D).

See the original guideline document for discussion of carditis, subcutaneous nodules and erythema marginatum.

Clinical Features of Acute Rheumatic Fever: Minor Manifestations

Fever

As there are no recent data relating to fever in low-risk populations, it is recommended that an oral, tympanic or rectal temperature greater than 38°C on admission, or documented with a reliable history during the current illness, should be considered as fever (level IV, Grade C).

Prolonged P-R Interval and Other Rhythm Abnormalities

An electrocardiogram (ECG) should be performed in all cases of suspected ARF (level IV, Grade C).

See the original guideline document for discussion of arthralgia and elevated acute-phase reactants.

Evidence of Group A Streptococcal Infection

Streptococcal Serology in High Incidence Populations

The values outlined in Table 3.6 (see the original guideline document) should be considered as normal values for both Aboriginal and non-Aboriginal populations (level IV, Grade C).

Syndromes That May Be Confused with Acute Rheumatic Fever

Post-streptococcal Reactive Arthritis

It is recommended that the diagnosis of post streptococcal reactive arthritis should rarely, if ever, be made in high-risk populations, and with caution in low-risk populations (Grade C). Diagnosed patients should receive secondary prophylaxis for at least 5 years (high-risk populations), or at least 1 year (low-risk populations) (Grade D). Echocardiography should be used to confirm the absence of valvular damage in all of these patients from both high- and low-risk populations before discontinuing secondary prophylaxis (Grade D).

Paediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS)

In high-risk populations, clinicians should rarely, if ever, make a diagnosis of PANDAS, and should rather err on the side of diagnosis of ARF and secondary prophylaxis (Grade D).

Echocardiography and Acute Rheumatic Fever

Echocardiography is more sensitive and specific for acute rheumatic carditis (level III-2) than auscultation, and today it is recommended that all patients with suspected or definite ARF should undergo echocardiography (Grade C).

Many case of ARF occur on the background of chronic RHD, and acute and chronic changes can co-exist. We currently recommend the following:

- In high-risk patients, pathological regurgitation of the mitral or aortic valve (in the absence of an alternative diagnosis, such as bicuspid aortic or mitral valve prolapse) is sufficient to fulfill the minimal echocardiographic criteria of acute carditis in the setting of suspected or proven ARF (Grade C).
- The presence of additional morphological changes to the mitral or aortic valve increases the confidence with which the diagnosis can be made (Grade C).
- Morphological changes of the mitral or aortic valve, in the absence of pathological valvular regurgitation, are not sufficient to diagnose acute rheumatic carditis. Such cases should be followed with repeat echocardiography after 4–6 weeks to detect evolving acute carditis (Grade D).

Valvulitis: Minimal Echocardiographic Criteria for Pathological Regurgitation

Regurgitation of the right-sided cardiac valves (tricuspid and pulmonary valve) is extremely rare without aortic or mitral valve involvement (level III-3). For this reason, a diagnosis of carditis should not be based on right-sided regurgitation alone (Grade C).

Subclinical Evidence of Rheumatic Valve Damage

The clinical course of subclinical carditis appears to be similar to that of mild carditis, with an audible murmur 5L (level III-2), and therefore, it is recommended that echocardiographically-detected valve damage (subclinical or otherwise) is included as a major manifestation of ARF in high-risk populations (Grade C).

Investigations

The recommended investigations in ARF are listed in the table below.

Table. Investigations in Suspected ARF Recommended for All Cases

Recommended for All Cases
White blood cell count
Erythrocyte sedimentation rate (ESR)
C-reactive protein (CRP)
Blood cultures, if febrile
Electrocardiogram (if prolonged P-R interval or other rhythm abnormality, repeat in 2 weeks and again at 2 months, if still abnormal)
Chest X-ray, if clinical or echocardiographic evidence of carditis
Echocardiogram (consider repeating after 1 month, if negative)
Throat swab (preferably before giving antibiotics): culture for group A streptococcus
Antistreptococcal serology: both ASO and anti-DNase B titres, if available (repeat 10–14 days later if first test not confirmatory)
Tests for Alternative Diagnoses, Depending on Clinical Features
Repeated blood cultures, if possible endocarditis
Joint aspirate (microscopy and culture) for possible septic arthritis
Copper, ceruloplasmin, antinuclear antibody, drug screen for choreiform movements
Serology and autoimmune

Management

Hospitalisation

See Table 3.11 in the original guideline document for priorities in managing ARF.

All patients with suspected ARF (first episode or recurrence) should be hospitalised as soon as possible after the onset of symptoms (Grade D).

Observation and General Hospital Care

Guidelines for general in-hospital care are provided in the table below (Grade D).

Table. Guidelines for General In-hospital Care

Nursing Recordings
Temperature, pulse, respiratory rate, blood pressure 4 times daily
Sleeping pulse (e.g., 0200 hours)
If pulse >100, apical heart rate

Diet
Free fluids (if no heart failure) Normal diet (limit extras) Early dietary advice if overweight and in heart failure, to avoid further weight gain Weekly weight
Bed Rest and General Care
Plan care to provide rest periods Provide age-appropriate activities Notify school teacher Involve family in care Prepare for discharge to primary care facility and follow up
If clinical carditis present (heart murmur, heart failure, pericardial effusion, valvular damage)
Document cardiac symptoms and signs Daily weight and fluid balance chart Diuretics, angiotensin-converting enzyme (ACE) inhibitors, digoxin if indicated; consider glucocorticoids Anticoagulation if atrial fibrillation present Cardiology opinion

Until the diagnosis is confirmed, it is recommended that joint pain be treated with paracetamol or codeine (Grade D).

Management of Probable Acute Rheumatic Fever

Patients with probable ARF may be managed in two ways (see Figure 3.1 in the original guideline document), according to the level of confidence with which the diagnosis is made (Grade D):

- Highly-suspected ARF: manage as for definite ARF
- Uncertain ARF: in patients from high-risk groups, administer 12 months of secondary prophylaxis initially, and reassess (including echocardiography) at that time. If there is no evidence of recurrent ARF, and no evidence of cardiac valvular damage on echocardiography, consider ceasing secondary prophylaxis. In such cases, the residual uncertainty should be discussed with the patient, and they should be encouraged to be particularly vigilant about treatment of sore throats, prevention and treatment of skin sores and early presentation with any symptoms of potentially recurrent ARF.

Treatment

Antibiotics

Controlled studies have failed to show that treating ARF with large doses of penicillin affects the outcome of rheumatic valvular lesions 1 year later. Despite this, most authorities recommend a course of penicillin, even if throat cultures are negative, to ensure eradication of streptococci that may persist in the upper respiratory tract (Grade D). This should be either a single injection of intramuscular benzathine penicillin G (BPG) (1,200,000 U or 600,000 U, if less than 20 kg) or a 10-day course of oral penicillin V (250 mg twice daily in children, 500 mg twice daily in adolescents and adults). Although a systematic review concluded that shorter-duration courses (3–6 days) of oral antibiotics may be an acceptable treatment of streptococcal pharyngitis in children in populations with low-risk ARF (level I, Grade A), the lack of studies in populations with high-risk ARF, a possible increased risk of late bacteriological recurrence in those receiving short courses and the absence of information about whether short course treatment can prevent ARF, suggest that this approach should not be recommended in populations at high risk of ARF (level I, Grade B). Similarly, it is not recommended as eradication treatment during an episode of ARF (Grade D).

Arthritis/Arthralgia

Anti-inflammatory therapy should be commenced in patients with arthritis or severe arthralgia as soon as the diagnosis of ARF has been confirmed (Grade B), but should be withheld if the diagnosis is not certain. In such cases, paracetamol or codeine should be used instead for pain relief (see Table 3.11 in the original guideline document).

Aspirin

It is recommended that children receiving aspirin during the influenza season (autumn/winter) also receive the influenza vaccine (Grade D).

Chorea

Although other causes should be excluded, in populations with endemic ARF, the vast majority of chorea presentations will be due to ARF, and neuroimaging is not needed routinely (Grade C).

Until more evidence is available, intravenous immunoglobulin (IVIG) is not recommended, except for severe chorea refractory to other treatments (level II/IV, Grade C).

Due to the small potential for liver toxicity with valproic acid, it is recommended that carbamazepine be used initially for severe chorea requiring treatment, and that valproic acid be considered for refractory cases (level III-2, Grade B).

Carditis/Heart Failure

The available evidence suggests that salicylates do not decrease the incidence of residual RHD (level IV). Therefore, salicylates are not recommended to treat carditis (Grade C).

Glucocorticoids may be considered for patients with heart failure in whom acute cardiac surgery is not indicated (Grade D).

There is little experience with beta blockers in heart failure, due to acute carditis, and their use is not recommended (Grade D).

Bed Rest

Patients with milder or no carditis should remain in bed only as long as necessary to manage other symptoms, such as joint pain (Grade D).

Secondary Prevention and Rheumatic Heart Disease Control Programs

Individual Approaches to Secondary Prevention

Secondary Prophylaxis

Antibiotic Regimens for Secondary Prophylaxis

It is recommended that 1,200,000 U of BPG should be used for secondary prophylaxis for all persons weighing 20 kg or more, and 600,000 U for those weighing less than 20 kg (level III-2, Grade B).

The use of four-weekly BPG is currently the treatment of choice, except in patients considered to be at 'high risk', for whom three-weekly administration is recommended. High-risk patient groups include:

- Those with moderate or severe carditis, or a history of valve surgery, who demonstrate good adherence to less frequent injections
- Those who have confirmed breakthrough ARF, despite full adherence to four-weekly BPG (see Table 4.2 in the original guideline document) (Grade D).

There are no data on the relative efficacies of these approaches, but the pharmacokinetic data suggest that prolonging the dosing interval beyond 4 weeks may increase the risk of breakthrough ARF. Therefore, monthly, rather than four-weekly administration of BPG, is an acceptable alternative, only if it is considered that the practicalities of monthly dosing will substantially improve adherence (Grade D).

Alternatives to Intramuscular Benzathine Penicillin G

Oral penicillin should be reserved for patients who experience bleeding problems following injection, and for those who refuse intramuscular BPG (level II, Grade B). If a patient is offered oral penicillin, the consequences of missed doses must be emphasised, and adherence carefully monitored (Grade D).

Penicillin Allergy

Before commencing penicillin treatment, patients should be carefully questioned about known allergies to penicillin and other beta-lactam antibiotics. If a confirmed, immediate and severe allergic reaction to penicillin is revealed, a non-beta-lactam antimicrobial (e.g. erythromycin) should be used instead (Grade D).

Secondary Prophylaxis in Pregnancy

As there is no evidence of teratogenicity, penicillin prophylaxis should continue for the duration of pregnancy for the prevention of recurrent ARF (Grade D).

Secondary Prophylaxis in Anticoagulated Patients

Intramuscular bleeding from BPG injections, used in conjunction with anticoagulation therapy in Australia, is rare. Thus, BPG injections should be continued for anticoagulated patients, unless there is evidence of uncontrolled bleeding, or the international normalised ratio (INR) is outside the defined therapeutic window (Grade D).

Duration of Secondary Prophylaxis

The appropriate duration of secondary prophylaxis is determined by age, persistence of environmental risk factors, time since the last episode of ARF and potential harm from recurrent ARF. Critical factors are outlined in Table 4.3 in the original guideline document. Based on these factors, the recommended duration of secondary prophylaxis is outlined in the table below (Grade D).

Table. Duration of Secondary Prophylaxis

Category	Definition of Category	Duration
All persons with ARF or RHD†		Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer).
Status after initial period elapsed:		
No RHD	No pathological mitral or aortic regurgitation, but may have minor morphological changes to mitral or aortic valves on echocardiography	Discontinue at that time‡
Mild RHD	Mild mitral or aortic regurgitation clinically and on echocardiography, with no clinical evidence of heart failure, and no evidence of cardiac chamber enlargement on echocardiography	Discontinue at that time
Moderate RHD	• Any valve lesion of moderate severity clinically (e.g., mild–moderate cardiomegaly and/or mild–moderate heart failure) or on echocardiography • Mild mitral regurgitation, together with mild aortic regurgitation clinically or on echocardiography • Mild or moderate mitral or aortic stenosis • Any pulmonary or tricuspid valve lesion coexisting with a left-sided valve lesion	Continued until 35 years of age
Severe RHD	• Any severe valve lesion clinically (e.g., moderate to severe cardiomegaly or heart failure) or on echocardiography • Any impending or previous cardiac valve surgery for RHD	Continued until age 40 years or longer*

†Patients >25 years of age who are diagnosed with RHD, without any documented history of prior ARF, should receive prophylaxis until the age of 35 years. At this time, they should be reassessed to determine whether prophylaxis should be continued.

‡Decisions to cease secondary prophylaxis should be based on clinical and echocardiographic assessment. *Risk of recurrence is extremely low in people aged >40 years. In some cases, for example, when the patient decides that they want to reduce even a minimal risk of recurrence, prophylaxis may be continued beyond the age of 40 years, or even for life.

Ceasing Secondary Prophylaxis

The duration of secondary prophylaxis should be based on individual needs, clinical features, social circumstances and the likelihood of ongoing exposure to GAS and further episodes of ARF.

It is reasonable to cease secondary prophylaxis at the age of 40, except when individual circumstances warrant continuation (e.g., when patients are keen to reduce even a small chance of a recurrence) (level IV, Grade C). Before stopping prophylaxis, recipients should be evaluated for symptomatic deterioration and the stability and severity of valve lesions. This should include echocardiographic assessment (Grade D).

Where limited echocardiography is available, preference should be given to patients with a history of moderate or greater carditis, a history of one or more ARF recurrences or clinical evidence of carditis (e.g., a murmur) (Grade D).

The date of cessation may be reviewed if there is a change in clinical or echocardiographic severity, a specialist recommendation or a recurrence of ARF (Grade D).

See the original guideline document for details on improving adherence to secondary prophylaxis.

Reducing the Pain of Benzathine Penicillin G Injections

The manufacturers of prepackaged syringes of BPG currently used in Australia for secondary prophylaxis do not recommend the addition of lignocaine or procaine penicillin (Grade D).

Direct application of pressure to the injection site has been shown to decrease pain of intramuscular injections. Other techniques that are easy to implement include warming refrigerated syringes to room temperature, ensuring that skin swabbed with alcohol is dry before injection and delivering the injection very slowly. As these measures are logical and benign, they are recommended, despite the lack of evidence (see Table below) (Grade D).

Table. Measures That May Reduce the Pain of BPG Injections

- Use a 21-gauge needle
- Warm syringe to room temperature immediately before using
- Allow alcohol from swab to dry before inserting needle
- Apply pressure with thumb for 10 sec before inserting needle
- Deliver injection very slowly (preferably over at least 2–3 min)
- Distract patient during injection (e.g., with conversation)

(The addition of 0.5–1 mL of 1% lignocaine is used elsewhere, but is not recommended with preloaded syringes currently available in Australia).

Prevention of Infective Endocarditis

Recommendations for procedures that require antibiotic prophylaxis are outlined in the table below and Table 4.8 in the original guideline document (Grade D).

In those without a history of iodine allergy, preprocedure use of antiseptic mouthwash is recommended (Grade C).

Irrespective of procedures, however, good oral health and dental hygiene are essential to reduce the likelihood of recurrent bacteraemia associated with day-to-day activities, and are likely to be important determinants of the degree and duration of bacteraemia associated with oral procedures. Patients should be supported in routine oral care, and regular dental reviews should be encouraged (Grade D).

Table. Procedures Requiring Endocarditis Prophylaxis in Patients with RHD

Dental
• Dental extractions • Periodontal procedures including surgery, subgingival scaling and root planning • Replanting avulsed teeth

- Other surgical procedures (e.g., implant placement, apioectomy)

Prophylaxis should be considered for the following if multiple procedures are being conducted, if prolonged or periodontal disease is evident:

- Periodontal probing; intraligamentary and intraosseous injections; surpagingival cleaning; rubber dam placement with clamps; restorative matrix band/strip placement; endodontics beyond the apical foramen; orthodontic bands; interdental wedges; subgingival placement of retraction cords, antibiotic fibres or strips

Respiratory Tract

Any procedure involving incision or biopsy of mucosa, such as:

- Tonsillectomy/adenoidectomy
- Flexible or rigid bronchoscopy (with incision or biopsy)
- Surgery of the bronchial, sinus, nasal or middle ear mucosa (including tympanoplasty)

Genitourinary and Gastrointestinal Tract

Any procedure where antibiotic prophylaxis is indicated for surgical reasons:

- Lithotripsy
- Vaginal delivery with prolonged labour
- Any genitourinary procedure in the presence of genitourinary infection
- Any gastrointestinal procedure in the presence of intra-abdominal infection

Other

- Incision and drainage of local abscess
- Surgical procedure through infected skin

Routine Review and Structured Care Planning

A structured care plan should be developed and recorded in the primary healthcare record of all persons with a history of ARF, or with established RHD (see Figure 4.1 in the original guideline document). Table 4.9 in the original guideline document lists recommended care plan schedules, which may be tailored to the needs of the individual (Grade D).

Dental Care

Routine dental care and appropriate oral hygiene is critically important in patients with a history of ARF and/or RHD. All patients should receive education about oral hygiene, and should be referred promptly for dental assessment and treatment when required. This is especially important prior to valvular surgery, when all oral/dental pathology should be investigated and treated accordingly (Grade D).

Organisational Approaches to Secondary Prevention

Primary prevention activities, aimed at preventing the first episode of ARF, should also be supported by the program. Specific elements are listed in the table below (Grade C).

Rheumatic Heart Disease Control Programs

Table. Recommended Elements of RHD Control Programs

- Commitment from national, regional and local services, particularly to ensure long-term funding and governance support
- An effective advisory committee that includes medical specialists, general practitioners, epidemiologists, nurses, public health practitioners, Aboriginal health service organisations and relevant community representatives
- A dedicated coordinating team
- An electronic patient register that contains data elements that support quality patient management, as well as any internal and external

reporting requirements
- Prioritisation of primary and secondary antibiotic prophylaxis delivered within the framework of primary healthcare
- Planning and advocacy for a stable supply of BPG, and establish plans for sustainable secondary prophylaxis in the event of supply reductions
- Development of the ability to find new cases of ARF and RHD and to assess and monitor the burden of disease
- A commitment to partnerships between clinicians and public health services in order to support the needs of people with ARF/RHD and the community
- Provision of education for health practitioners and health workers, and supported health education for the community, those with disease and their families
- Activities guided by locally relevant, evidence-based guidelines
- Legislation and/or regulations warranting the notification of ARF/RHD which is supported by public health surveillance activities at the state or territory level
- A priority system that ensures services are delivered to those at highest risk
- A mechanism for monitoring delivery of secondary prophylaxis and ongoing care
- Evaluation of patient management and program activities

Public Health Approaches to Acute Rheumatic Fever and Rheumatic Heart Disease Control

Surveillance

Ideally, active surveillance should be used to augment passive surveillance (Grade D).

Screening for Rheumatic Heart Disease

Currently, it is recommended that RHD control programs should coordinate screening to detect previously undiagnosed RHD in high-risk populations, wherever this is practical (Grade D). Although RHD prevalence is highest in adults, they are difficult to screen. It is recommended that screening rather focuses on school-aged children (Grade D).

Recent studies suggest that auscultation is poorly sensitive and specific, and that echocardiographic screening may be the best method. The availability of portable echocardiography, and the ability to perform a limited assessment of the mitral and aortic valve in only 5–10 min, make echocardiographic screening feasible. Where echocardiography is not available to review all children with murmurs, a highly experienced auscultator could select all children with non-innocent murmurs for echocardiography (Grade D).

Evaluating Rheumatic Heart Disease Control Programs

As has been highlighted throughout the developing world, the availability of and support for routine primary healthcare is essential in controlling ARF/RHD.

Indicators used to evaluate ARF/RHD control programs should be relevant, structured, measurable, routinely available and affordable. In particular, they should not overburden primary healthcare providers, and should lead to improved clinical results. A list of suggested indicators is provided in Appendix 3 of the original guideline document (Grade D).

Diagnosis and Management of Rheumatic Heart Disease

Background and Management Principles

Access to Oral Healthcare

Oral health assessment is part of routine management of RHD. It is recommended that all patients with RHD (regardless of severity) undergo annual oral health review (Grade D).

Access to Echocardiography

Many patients with RHD do not have a documented history of ARF, and it may be difficult to judge their symptomatic status by standard clinical criteria (e.g., New York Heart Association Functional Class [NYHA FC]) because of communication difficulties and cultural barriers. For example, many Aboriginal and Torres Strait Islander patients, especially those from remote communities, report few symptoms, even in the presence of advanced valvular disease (Grade D).

All patients with murmurs suggestive of possible valve disease, or a history of ARF, require echocardiography (Grade D).

Access to Cardiothoracic and Interventional Cardiology Services

Clinical symptoms and the nature of the predominant lesion should dictate the medical management and timing of cardiac intervention (Grade D).

Echocardiographic Criteria for Rheumatic Heart Disease

The WHF recommends two echocardiographic categories of RHD in individuals ≤20 years of age: 'definite RHD' and 'borderline RHD', based on evidence derived from numerous studies (level III-2, IV, Grade C). The borderline RHD category was established to improve the sensitivity of the test for individuals from regions with a high prevalence of RHD (i.e., high-risk populations), and who, due to their young age, may not have had sufficient time to develop the full echocardiographic manifestations of RHD. The borderline category is not applicable to patients who are considered to be at low risk of RHD, and therefore, those with a low pretest probability (Grade D). In individuals who are aged over 20 years, minor age-related or degenerative changes may overlap with what is defined as borderline RHD on echocardiography. Hence, the use of the borderline RHD category is not advised in adults beyond 20 years of age (level 4, Grade C).

Criteria for pathological regurgitation and morphological features of RHD are detailed in Tables 5.2 and 5.3 of the original guideline document. Trivial regurgitation of the mitral valve, and even of the aortic (that does not meet all four criteria for pathological regurgitation), is common and should be considered normal or physiological (level III-2, Grade C) (see Tables 5.1 and 5.2 in the original guideline document). The same can be said for isolated morphological changes, such as valvular thickening, that occurs without pathological stenosis or regurgitation (level III-3, Grade C).

Mitral Regurgitation (MR)

Medical Management

The absence of increased afterload in MR (instead, there is a low resistance leak into the left atrium) suggests that vasodilator therapy is unlikely to be beneficial in improving outcome. Therefore, this drug therapy is not recommended in the medical management of MR, unless there is associated heart failure, LV dysfunction or hypertension (level IV, Grade C).

In asymptomatic or mildly-symptomatic patients with moderate or more severe MR, echocardiography should be performed at least every 6–12 months (Grade D).

Surgical Management

Indications for Surgery

Patients who are symptomatic, with moderate to severe MR, should be automatically referred for surgical management (level III-2, Grade C). Patients who develop very enlarged hearts (adults left ventricular end-systolic dimension [LVESD] ≥40 mm; a critical LV end-systolic dimension has not been identified in children) or impaired systolic function (ejection fraction [EF] <60%) have an increased surgical risk, less likelihood of restoring normal systolic function and increased risk of late heart failure and death (level III-2). This also applies to those with significant pulmonary hypertension (>50 mmHg) and preoperative AF (level III-2). In addition, the presence of AF for more than 1 year is a predictor of persistence of AF after successful valve surgery. Therefore, it is recommended that in the setting of severe chronic MR, patients should be recommended for surgery once those parameters are approached, rather than reached, regardless of symptomatic status (level III-2, Grade C) (see Table 5.5 in the original guideline document), especially in children and young people.

Mitral Stenosis (MS)

Atrial Fibrillation (AF)

Patients with MS and chronic or paroxysmal AF should receive long-term prophylactic anticoagulation with warfarin (level III-3, Grade C). However, left atrial thrombus can occur in MS, even when sinus rhythm is present, due to left atrial dilatation, low blood velocity and disorganised blood flow. Therefore, prophylactic anticoagulation should also be considered for patients with MS, a large left atrium and sinus rhythm (Grade D).

When new-onset AF is symptomatic, consideration should be given to direct current cardioversion to restore sinus rhythm (Grade B).

Percutaneous Balloon Mitral Valvuloplasty (PBMV)

The treatment of choice for dominant or pure MS is percutaneous balloon mitral valvuloplasty (PBMV) (level III, Grade B).

Indications for Balloon Valvuloplasty

The indication for PBMV is progressive exertional dyspnoea (NYHA FC II, III or IV), associated with documented evidence of moderate or

severe MS (mitral orifice area <1.5 cm^2) (Grade B).

Surgical Management

PBMV has largely replaced surgical mitral commissuroplasty and commissurotomy. In the relatively few patients who are not suitable for PBMV, every effort should be made to repair the mitral valve, rather than replace it, if patients are in sinus rhythm (Grade D)

See Table 5.6 in the original guideline document for the key points in the management of rheumatic mitral stenosis.

Aortic Regurgitation

Medical Management

Until there are more trial data, vasodilator therapy with nifedipine or ACE inhibitors is still recommended for asymptomatic or mildly-symptomatic patients with preserved systolic function and moderate or greater degrees of AR (level III-3, Grade C) especially when systolic hypertension is present.

Surgical Management

Indication for Surgery

Symptomatic patients with moderate/severe AR should be referred for surgery (level III-2, Grade B)

Patients with equivocal symptoms may undergo exercise testing, which is useful in assessing functional capacity and symptomatic response (level IV, Grade C).

If serial echocardiography shows that the LVESD is approaching 55 mm, or the LVEF is <55%, these patients should be referred for aortic valve surgery (Grade C).

Recommendations

A careful preoperative assessment of the likelihood of medication adherence, especially warfarin, is essential in determining the choice of valve surgery. If stable anticoagulation is unlikely to be achieved, serious consideration should be given to the use of an aortic bioprosthesis. Patients who demonstrate good adherence with medications are suitable for replacement with the newer bileaflet mechanical valve prosthesis, as they have the best long-term durability and highest freedom from reoperation. However, in young female patients, every effort must be made to avoid a mechanical prosthesis, because of the significant risk to mother and fetus posed by anticoagulation during pregnancy.

Aortic Stenosis (AS)

Indications for Surgery

Aortic valve replacement is a definitive therapy for symptomatic AS (level III-2, Grade B). It should be performed in all patients with significant gradients and a reduced valve orifice (mean gradient >40-50 mm, aortic valve orifice <1 cm^2), once they develop exertional symptoms. It should also be considered in patients with significant LV dysfunction, but with a lower aortic gradient.

Rheumatic Tricuspid Valve Disease

Surgical Management

Surgical management of rheumatic tricuspid valve disease may be challenging, and is frequently associated with left-sided rheumatic valve disease, especially mitral valve disease. In most cases, the tricuspid valve can be repaired without the need for prosthetic valve replacement. The alternative to tricuspid valve repair is valve replacement.

Pregnancy in Patients with Rheumatic Heart Disease

Mitral/Aortic Regurgitation

Hydralazine and nitrates, or dihydropyridine calcium channel blockers (e.g., nifedipine), hydralazine or nitrates, should be used if vasodilator therapy is needed (level IV, Grade C).

Mechanical Prosthetic Valves: Management of Anticoagulation Therapy

Patients with mechanical prosthetic valves should be given appropriate contraceptive advice to avoid unplanned pregnancy, and counselled about

the risks to mother and fetus with pregnancy (Grade D).

Given the maternal risks, the option of not continuing the pregnancy should be discussed. Some women may choose to increase their own risk for the benefit of their baby, and such decisions need to be respected, provided it is assured that she has a full understanding of the matter. After the patient agrees to the use of an anticoagulant regimen, written consent should be obtained, or the decision fully documented in the patient's health record (Grade D).

Recommendations for Anticoagulation in Pregnancy for Patients with Mechanical Valves

There are limited published data available on anticoagulant options, and no randomised comparative studies have been or are likely to be performed. There is a choice of three different anticoagulant regimens during pregnancy for patients with mechanical prostheses (level IV, Grade C): LMWH (enoxaparin) throughout pregnancy, LMWH/warfarin, and warfarin throughout pregnancy (especially older prostheses).

Management of Delivery

Breastfeeding can be encouraged in women taking anticoagulants, as heparin is not secreted in breast milk; the amount of warfarin in breast milk is low and has been shown to have no effect on neonatal prothrombin time (Grade C).

Definitions:

Grade of Recommendation

A	Rich body of high-quality randomised, controlled trial (RCT) data
B	Limited body of RCT data or high-quality non-RCT data
C	Limited evidence
D	No evidence available; panel consensus judgement

Levels of Evidence

Level of Evidence	Study Design
I	Evidence obtained from a systematic review of all relevant randomised, controlled trials (RCTs)
II	Evidence obtained from at least one properly designed RCT
III-1	Evidence obtained from well-designed pseudo RCT (alternate allocation or some other method)
III-2	Evidence obtained from comparative studies, with concurrent controls and allocation not randomised (cohort studies), case-control studies, or interrupted time series with a control group
III-3	Evidence obtained from comparative studies with historical control, two or more single-arm studies or interrupted time series without a parallel control group
IV	Evidence obtained from case series, either post-test or pretest and post-test

Note: The levels of evidence are adapted from the National Health and Medical Research Council levels of evidence for clinical interventions and the US National Institutes of Health clinical guidelines (details can be found at www.nhlbi.nih.gov/guidelines/obesity/ob_home.htm).

Clinical Algorithm(s)

The following clinical algorithms are provided in the original guideline document:

- Outline of structure for preventive strategies for GAS pharyngeal colonisation and pharyngitis
- Management of probable ARF

- Recommended routine review and structured care planning
- Timing of surgery for severe mitral regurgitation
- Timing of surgery for mitral stenosis
- Timing of surgery for aortic regurgitation
- Timing of surgery for aortic stenosis

Scope

Disease/Condition(s)

- Acute rheumatic fever (ARF)
- Rheumatic heart disease (RHD)

Guideline Category

Diagnosis

Management

Prevention

Treatment

Clinical Specialty

Cardiology

Infectious Diseases

Internal Medicine

Pediatrics

Rheumatology

Thoracic Surgery

Intended Users

Advanced Practice Nurses

Allied Health Personnel

Health Care Providers

Nurses

Physician Assistants

Physicians

Public Health Departments

Guideline Objective(s)

- To improve health outcomes for people with (or at risk of developing) acute rheumatic fever (ARF) and rheumatic heart disease (RHD), and to encourage the use of appropriate resources
- To identify the standard of care, including preventive care, which should be available to all people
- To identify areas where current management strategies may not be in line with available evidence
- To ensure, in the interests of equity, that high-risk populations receive the same standard of care as that available to other Australians

Target Population

Patients with or at risk of developing acute rheumatic fever or rheumatic heart disease

Interventions and Practices Considered

Assessment/Diagnosis

1. Medical history and physical examination
2. Echocardiography
3. Electrocardiography
4. Laboratory tests (C reactive protein, erythrocyte sedimentation rate, white blood cell count, blood cultures, antistreptococcal serology)
5. Chest X-ray
6. Throat swab and culture for group A streptococcus
7. Exercise test

Management/Treatment

1. Antibiotics (intramuscular benzathine penicillin G, oral penicillin V or oral erythromycin)
2. Management of arthritis/arthralgia (paracetamol, codeine, aspirin, naproxen or ibuprofen)
3. Influenza vaccination
4. Management of chorea (carbamazepine or valproic acid, immunoglobulin in cases refractory to other treatment)
5. Management of fever (salicylates, paracetamol)
6. Management of carditis/heart failure (bed rest, echocardiogram, diuretics/fluid restriction, glucocorticoids, salicylates, angiotensin-converting enzyme [ACE] inhibitors, digoxin, valve surgery)
7. Prevention (primary, secondary prophylaxis)
8. Hospitalisation
    - Monitoring (temperature, pulse, respiratory rate)
    - Diet
    - Bed rest/general care
9. Management of clinical carditis (documentation of cardiac symptoms and signs, weight and fluid balance chart, diuretics, ACE inhibitors, digoxin, glucocorticoids, anticoagulants, referral to cardiology)
10. Routine review and structured care planning
11. Management of mitral regurgitation (surgery)
12. Management of mitral stenosis (diuretics, anticoagulants, cardioversion, percutaneous balloon mitral valvuloplasty, surgery)
13. Management of aortic regurgitation (vasodilator therapy, ACE inhibitors, surgery)
14. Management of aortic stenosis (surgery)
15. Management of rheumatic tricuspid valve disease (diuretics, surgery)
16. Management of acute rheumatic fever/rheumatic heart disease in pregnancy

Major Outcomes Considered

- Sensitivity and specificity of diagnostic tests
- Adherence to secondary prophylaxis
- Rates of further attacks of acute rheumatic fever (ARF), cardiac damage, and premature death
- Rates of surgery because of rheumatic heart disease (RHD)
- Incidence of antibiotic resistance with Group A streptococcal prophylaxis

- Time from diagnosis of ARF to hospitalisation and treatment
- Incidence of fetal complications in pregnant women receiving secondary prophylaxis
- Duration of secondary prophylaxis
- Incidence of infective carditis in patients with RHD

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

A comprehensive and systematic literature review of all publications in acute rheumatic fever (ARF) and rheumatic heart disease (RHD) since 2004 was undertaken. The titles and abstracts of these articles were scanned to select those that might offer new information around the four sections in these guidelines (primordial and primary prevention, ARF diagnosis and management, secondary prevention and control programs and management of RHD).

Databases Searched

The following databases were searched:

- The Cochrane Library of Systematic Reviews and Cochrane Databases
- Ovid Medline
- EMBASE
- PUBMED
- PUBMED Clinical trials
- Scirus Search (deep web)
- Public Library of Science (PLoS - free access database)

Search Terms

The following search terms were used:

- Acute rheumatic fever
- Rheumatic heart disease
- Diagnosis
- Management
- Treatment
- Acute rheumatic fever AND diagnosis OR management OR treatment
- Rheumatic heart disease AND diagnosis OR management OR treatment
- Chorea
- Rheumatic chorea
- Sydenham's chorea
- Mitral stenosis
- Mitral regurgitation (rheumatic only)
- Percutaneous mitral valvuloplasty
- Pregnancy and rheumatic heart disease
- Pregnancy and valvular heart disease

Limits

- Date range – 2004 to 'current' (or 31 May 2010)
- Articles in English
- Studies with humans

Other searches were run to ensure that all the terms in the 2006 guidelines document and those specifically identified by the expert group were included. These were:

- Echocardiography
- Serology (new data, new ranges)
- Post-streptococcal sequelae
- Post-streptococcal reactive arthritis
- PANDAS
- Subclinical carditis
- Pregnancy

Number of Source Documents

406 unique references were sent to the expert group for review.

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Levels of Evidence

Level of Evidence	Study Design
I	Evidence obtained from a systematic review of all relevant randomised, controlled trials (RCTs)
II	Evidence obtained from at least one properly designed RCT
III-1	Evidence obtained from well-designed pseudo RCT (alternate allocation or some other method)
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III-3	Evidence obtained from comparative studies with historical control, two or more single-arm studies or interrupted time series without a parallel control group
IV	Evidence obtained from case series, either post-test or pretest and post-test

Note: The levels of evidence are adapted from the National Health and Medical Research Council levels of evidence for clinical interventions and the US National Institutes of Health clinical guidelines (details can be found at www.nhlbi.nih.gov/guidelines/obesity/ob_home.htm).

Methods Used to Analyze the Evidence

Systematic Review

Description of the Methods Used to Analyze the Evidence

The full text of articles found by the systematic review was reviewed by members of the writing group for the relevant section.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

This document was developed by Rheumatic Heart Disease Australia (RHDAustralia), in collaboration with the Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand (CSANZ). A revision of the original evidence-based review was undertaken by a core multidisciplinary writing group (listed on page 2 of the original guideline document).

- The core writing group prepared an updated version of the existing content, including the new evidence, and an additional, new section on primordial and primary prevention was developed.
- Selected reviewers with clinical and public health experience in ARF and RHD then reviewed each chapter, and their suggestions were incorporated into a second draft.
- A number of additional amendments were made following ongoing discussion.

Rating Scheme for the Strength of the Recommendations

Grade of Recommendation

A	Rich body of high-quality randomised, controlled trial (RCT) data
B	Limited body of RCT data or high-quality non-RCT data
C	Limited evidence
D	No evidence available; panel consensus judgement

Note: The levels of evidence and grades of recommendations are adapted from the National Health and Medical Research Council levels of evidence for clinical interventions and the US National Institutes of Health clinical guidelines (details can be found at www.nhlbi.nih.gov/guidelines/obesity/ob_home.htm).

Cost Analysis

- Secondary prophylaxis with benzathine penicillin G (BPG) is the only rheumatic heart disease (RHD) control strategy shown to be clinically effective and cost-effective at both community and population levels. Randomised, controlled trials (RCT) have shown that regular administration is required to prevent recurrent acute rheumatic fever (ARF).
- RHD control programs aim to improve the delivery of secondary prophylaxis, the most cost-effective approach to RHD control. This approach has been estimated to cost less than half that of tertiary services (including cardiac surgery), and less than one-seventh that of primary prophylaxis. The management of chronic RHD has been estimated to consume up to 70% of the total national ARF/RHD budget for New Zealand. A recent review on RHD in the Pacific region found that the cost of tertiary-level surgical intervention for one patient is equivalent to the annual running costs for a national RHD control program in Pacific countries with small populations. There is little doubt that much of this expenditure could be prevented with targeted and coordinated secondary prevention programs.
- While it is possible to treat all cases of pharyngitis with antibiotics, this would expose a significant proportion of patients to unnecessary treatment, as only 20%–40% of pharyngitis episodes are associated with group A streptococcal (GAS) infection; the remainder are caused by viruses or by bacteria for which antibiotic treatment is not recommended. Moreover, such an approach would require substantial resources and expose clients to unwarranted inconvenience and risk, while increasing the possibility of antibiotic resistance. However, some treatment guidelines do suggest that people identified as being from populations at high risk of ARF, or who have established RHD, but are not currently receiving secondary antibiotic prophylaxis, should be treated with antibiotics if they develop pharyngitis, irrespective of other clinical features, and before confirmatory testing for GAS is available (see Table 2.2 in the original guideline document). While empirically attractive, there is no clear evidence that such an approach is a safe or cost-effective way to reduce the incidence of ARF.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

The final draft was endorsed by of the main authors and reviewers at an editorial meeting in October 2011. The document was then distributed to a range of stakeholders for endorsement (see the "Guideline Endorser[s]" field).

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type evidence supporting each recommendation is specifically stated (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Accurate diagnosis and appropriate prevention and management of acute rheumatic fever and rheumatic heart disease

Potential Harms

- Side effects of drug therapy, including drug interactions and allergy
- Complications of surgery and invasive procedures
- Maternal and/or fetal complications from warfarin and heparin

Contraindications

Contraindications

- A large left atrial thrombus is a contraindication to percutaneous balloon mitral valvuloplasty (PBMV).
- Diltiazem, angiotensin receptor antagonists and angiotensin-converting enzyme (ACE) inhibitors are contraindicated during pregnancy.
- Valproic acid should be avoided in women who are or who may be pregnant, because of the potential for damage to the fetus.
- In young female patients, every effort must be made to avoid a mechanical prosthesis, because of the significant risk to mother and fetus posed by anticoagulation during pregnancy.

Qualifying Statements

Qualifying Statements

- This revised national guideline provides a general framework, and should not override good clinical judgement. Treatment should take into account the patient's co-morbidities, drug tolerance, lifestyle, living circumstances, cultural sensibilities and wishes. When prescribing medication, clinicians should observe usual contraindications, be mindful of potential adverse drug interactions and allergies and monitor

responses and review patients regularly.

- This publication was funded by the Australian Government Department of Health and Ageing. The views expressed are those of the authors and do not necessarily reflect those of the Australian Government Department of Health and Ageing. The Commonwealth of Australia does not warrant or represent that the information contained in this publication is accurate, current or complete. The Commonwealth of Australia does not accept any legal liability or responsibility for any loss, damages, costs or expenses incurred by the use of, or reliance on, or interpretation of, the information contained in this publication.
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Implementation of the Guideline

Description of Implementation Strategy

The implementation of guidelines for chronic rheumatic heart disease (RHD) has major implications for the healthcare services of Aboriginal people and Torres Strait Islanders, especially in rural and remote regions. In addition to access to culturally-appropriate primary care services, best practice for RHD requires:

- Secondary prevention with penicillin prophylaxis
- Adequate monitoring of anticoagulation therapy in patients with atrial fibrillation (AF) and/or mechanical prosthetic valves
- Access to oral healthcare
- Access to echocardiography
- Access to a specialist physician, paediatrician and/or cardiologist, preferably the same specialist, for regular follow up visits
- Access to cardiothoracic and interventional cardiology services

Implementation Tools

Audit Criteria/Indicators

Clinical Algorithm

Mobile Device Resources

Patient Resources

Pocket Guide/Reference Cards

Quick Reference Guides/Physician Guides

Resources

Staff Training/Competency Material

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

Staying Healthy

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

RHDAustralia (ARF/RHD writing group), National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand. Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). Casuarina (Australia): RHDAustralia; 2012. 134 p. [459 references]

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2006 Jun (revised 2012)

Guideline Developer(s)

Cardiac Society of Australia and New Zealand - Disease Specific Society

National Heart Foundation of Australia - Disease Specific Society

RHDAustralia - Disease Specific Society

Source(s) of Funding

This publication was funded by the Australian Government Department of Health and Ageing. The views expressed are those of the authors and do not necessarily reflect those of the Australian Government Department of Health and Ageing.

Guideline Committee

ARF/RHD Writing Group

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Financial Disclosures/Conflicts of Interest

Not stated

Guideline Endorser(s)

Australasian Society for Infectious Diseases - Disease Specific Society

Australian and New Zealand Society of Cardiac and Thoracic Surgeons - Medical Specialty Society

Australian College of Rural and Remote Medicine - Professional Association

Australian Indigenous Doctors Association - Professional Association

Cardiac Society of Australia and New Zealand - Disease Specific Society

Council of Remote Area Nurses of Australia Inc. - Professional Association

Internal Medicine Society of Australia and New Zealand - Medical Specialty Society

National Aboriginal Community Controlled Health Organisation - National Government Agency [Non-U.S.]

National Heart Foundation of Australia - Disease Specific Society

Public Health Association of Australia - National Government Agency [Non-U.S.]

Royal Australasian College of Physicians - Professional Association

Royal Australian College of General Practitioners - Professional Association

Society of Obstetric Medicine of Australia and New Zealand - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: ARF/RHD Guideline Development Working Group of the National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand. Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia: an evidence-based review. Sydney (Australia): National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand; 2006 Jun. 84 p.

Guideline Availability

Electronic copies: Available in Portable Document Format (PDF) from the RHDAustralia Web site .

Availability of Companion Documents

The following are available:

- Primary prevention of acute rheumatic fever. Quick reference guide. 2012. 3 p. Electronic copies: Available in Portable Document Format (PDF) from the RHDAustralia Web site .
- Diagnosis of acute rheumatic fever. Quick reference guide. 2012. 7 p. Electronic copies: Available in PDF from the RHDAustralia Web site .
- Management of acute rheumatic fever. Quick reference guide. 2012. 7 p. Electronic copies: Available in PDF from the RHDAustralia Web site .
- Secondary prevention of acute rheumatic fever. Quick reference guide. 2012. 9 p. Electronic copies: Available in PDF from the RHDAustralia Web site .
- Management of rheumatic heart disease. Quick reference guide. 2012. 9 p. Electronic copies: Available in PDF from the RHDAustralia Web site .
- Rheumatic heart disease in pregnancy. Quick reference guide. 2012. 4 p. Electronic copies: Available in PDF from the RHDAustralia Web site .
- Rheumatic heart disease control programs. Quick reference guide. 2012. 7 p. Electronic copies: Available in PDF from the RHDAustralia Web site .
- Red flag tool for recognition of acute rheumatic fever (ARF). Pocket guide. 2012. 2 p. Electronic copies: Available in PDF from the RHDAustralia Web site .
- 10 things you should know about rheumatic fever. 2012. 1 p. Electronic copies: Available in PDF from the RHDAustralia Web site .

In addition, an app for mobile devices is available from the RHDAustralia Web site . E-learning modules are also available from the RHDAustralia Web site . Performance indicators and the recommended dataset for acute rheumatic fever/rheumatic heart disease are available in the appendices to the original guideline document .

Patient Resources

The following is available:

- What is acute rheumatic fever and rheumatic heart disease? Brochure. 2012. Electronic copies: Available in Portable Document Format from the RHDAustralia Web site .

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

NGC Status

This NGC summary was completed by ECRI on April 9, 2007. The information was verified by the guideline developer on June 27, 2007. This summary was updated by ECRI Institute on September 7, 2007 following the revised U.S. Food and Drug Administration (FDA) advisory on Coumadin (warfarin). This summary was updated by ECRI Institute on March 14, 2008 following the updated FDA advisory on heparin sodium injection. This summary was updated by ECRI Institute on December 26, 2008 following the FDA advisory on Innohep (tinzaparin). This summary was updated by ECRI Institute on November 12, 2010 following the U.S. Food and Drug Administration (FDA) advisory on Afluria (influenza virus vaccine). This summary was updated by ECRI Institute on March 16, 2011 following the U.S. Food and Drug Administration advisory on acetaminophen-containing prescription products. This NGC summary was updated by ECRI Institute on February 5, 2013. This summary was updated by ECRI Institute on October 28, 2013 following the U.S. Food and Drug Administration advisory on Acetaminophen. This summary was updated by ECRI Institute on March 10, 2014 following the U.S. Food and Drug Administration advisory on Low Molecular Weight Heparins. This summary was updated by ECRI Institute on September 18, 2015 following the U.S. Food and Drug Administration advisory on non-aspirin nonsteroidal anti-inflammatory drugs (NSAIDs). This summary was updated by ECRI Institute on June 2, 2016 following the U.S. Food and Drug Administration advisory on Opioid pain medicines.

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